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Case Report

Celiac Artery Compression Syndrome (CACS): A Case Report and Review of the Literature

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Abstract Celiac artery compression syndrome (CACS), also known as Dunbar syndrome or median arcuate ligament syndrome (MALS), is a condition that can result from a fibrous ligament, the median arcuate ligament, passing superior to the celiac axis causing extrinsic compression or entrapment of sympathetic neural fibers near its takeoff from the abdominal aorta. This uncommon disorder is characterized clinically by the triad of postprandial abdominal pain, weight loss, and sometimes abdominal bruit. In the literature, most cases of CACS are from Western countries. The diagnosis is generally considered after more common conditions have been ruled out. In particular, an extensive evaluation of GI tract and biliary system have been done in an effort to identify the cause of pain. For patients in whom celiac artery compression is suspected, the diagnosis requires vascular imaging with a respiratory maneuver to confirm CACS. The author reported a 27-year-old woman with CACS, along with results of cross-sectional CTA, 3D reconstruction, combined with intra-operative duplex ultrasound of the celiac artery to demonstrate stenosis, and an attempt at decompression by aggressive resection of median arcuate ligament and nerve fibers.

Keywords: Celiac artery compression syndrome, Median arcuate ligament, Dunbar syndrome

Introduction

Celiac artery compression syndrome (CACS), also called the median arcuate ligament syndrome (MALS)[1] or Dunbar syndrome[2], is defined as a chronic, recurrent abdominal pain related to compression of the celiac artery by the median arcuate ligament, which is a fibrous arch that traverses the aorta and bridges the crura of the diaphragm. The median arcuate ligament usually comes into contact with the celiac artery and the aorta at the level between T11 and L1, at the celiac artery origin, which is prone to compression during expiration (Figure 1).

CACS was firstly observed by Benjamin Lipshutz in 1917 and median arcuate ligament syndrome was described by Pekka-Tapani Harjola in 1963. The first clinical study of MALS was carried out by J. David Dunbar and Samuel Marable in 1965. MALS has also been called the Harjola-Marable syndrome and the Marable syndrome. Subsequently, many reports addressing MALS have been published. The diagnosis of this uncommon disorder can be done with cross-sectional abdominal CT or MRA (magnetic resonance angiography) plus three-dimensional reconstruction combined with a duplex ultrasonography study during appropriate respiratory maneuvers.

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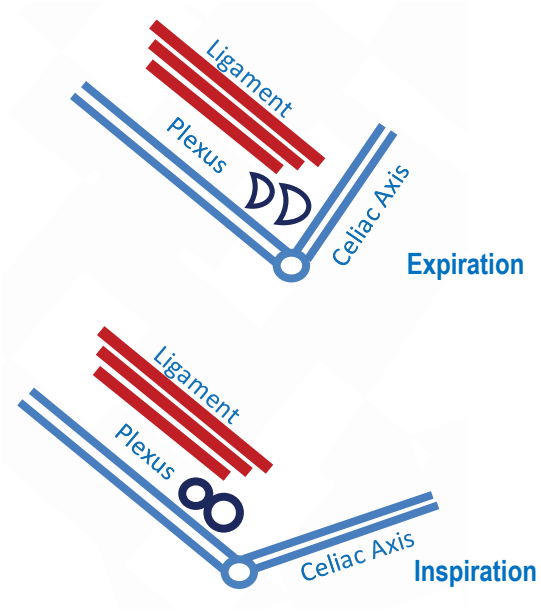

Figure 1 Celiac axis movement during normal respiration

The treatment of celiac artery decompression is generally by surgical division of median arcuate ligament combined with removal of the celiac ganglia. In the present study, the author reported a case of celiac artery compression by the median arcuate ligament in 27-year-old woman with symptoms of intermittent intestinal angina related to meals who experienced weight loss and was successfully treated by using a surgical approach along with intra-operative duplex ultrasonographic study.

Case Report

A 27-year-old woman experienced, for 3 months, epigastric pain that was aggravated by food intake. The pain continued after meals and increased severity to 7/10 on the visual analog scale (VAS), which sometimes radiated to the back. She also reported a weight loss of 4 kilograms over 4 weeks period, with symptoms of constipation. Her BMI was 18.37 kg/m^2. She was treated with proton-pump inhibiters (PPIs) for more over 2 weeks, without relief. She denied smoking, or alcohol and non-steroidal anti-inflammatory drugs (NSAIDs) use.

A diagnostic esophagogastroduodenoscopy (EGD) was performed, which showed no ulcer or evidence of *H.pylori* infection. An abdominal ultrasound showed normal hepato-biliary system and pancreas. She had no underlying diseases including psychiatric disorder. Physical examination revealed mild tenderness at the epigastriun without guarding and no audible abdominal bruit was detected. Routine laboratory blood tests and urine examination were normal. Abdominal and chest X-ray showed no abnormalities. These clinical signs and results of investigational studies were out of proportion to her symptoms, raising the suspicion of visceral ischemia. Further evaluation with computerized tomography angiography (CTA) revealed acute angulation and focal narrowing of the celiac axis with post-stenotic dilatation and demonstrating a hook-like appearance on the sagittal plane of CTA (Figure 2). Surgical intervention was planned for the patient.

Trans-abdominal duplex ultrasonography was done after general anesthesia, to measure the blood flow through celiac artery, and found that the peak systolic velocity (PSV) was 230 cm/sec (Figure 3), with no plaque identified. This suggests celiac artery stenosis associated with CACS or MALS. The patient therefore underwent open surgery for celiac artery decompression by dividing the median arcuate ligament. Surgery was performed through an upper midline abdominal incision. A Thompson's retractor was placed at the upper costal margin to elevate the left lobe of the liver and body of stomach.

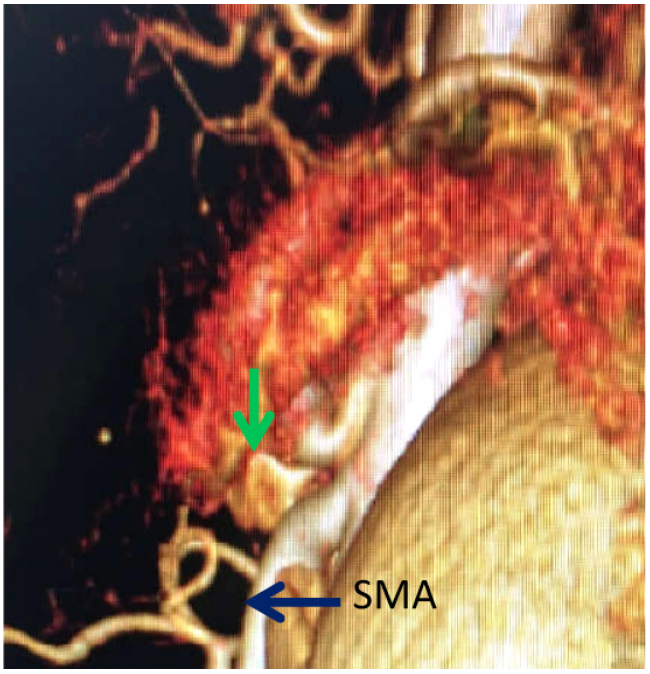

Figure 2 Lateral view of a CTA of the abdominal aorta and visceral takeoff, with a 3-D reconstruction, demonstrating of the celiac axis compression, acute angulation, post stenotic dilatation and hook-like appearance (green arrow) of the celiac artery.

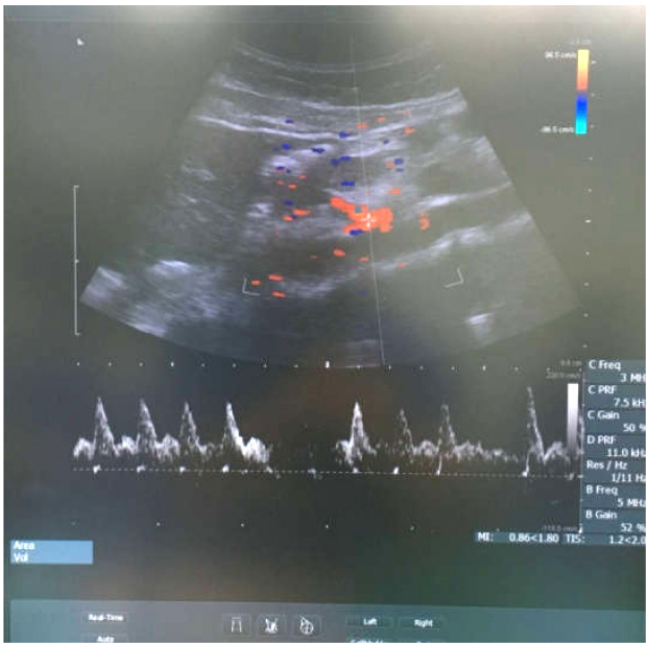

Figure 3 Duplex ultrasound of the abdominal aorta, showing a celiac artery peak-systolic velocity of 230 cm/sec, suggesting CACS.

The lesser sac was entered via the gastrocolic ligament, the gastrohepatic ligament was divided after mobilization of the left lobe of liver medially, and the aortic pulse just above the superior pancreatic border could be felt. By this method, the splenic artery, common hepatic artery, and left gastric artery were identified and exposed at their origin down to celiac trunk and abdominal aorta.

The short celiac trunk, which was compressed by muscular fibers from the diaphragm, was well identified (Figure 4). Aggressive resection of the median arcuate ligament and nerve fibers was done, the diaphragmatic crura were mobilized away from the celiac artery, exposing the anterior abdominal aorta up to 4 cm (Figure 5).

Confirmation of the completeness in celiac artery decompression was done by visual inspection and assessing celiac flow with intra-operative intra-operative duplex ultrasound to determine the adequacy of treatment, in which the peak-systolic velocity was 120 cm/sec (Figure 6).

On the first day after the operation, the patient was given oral diet, advancing as tolerated, and discharged home after a hospitalization of 3 days. The patient was followed post-operatively at 1 month to evaluate the incision site, dietary intake, recurrent symptoms, and a duplex scanning of the celiac artery was done at 6 months, all of which were normal.

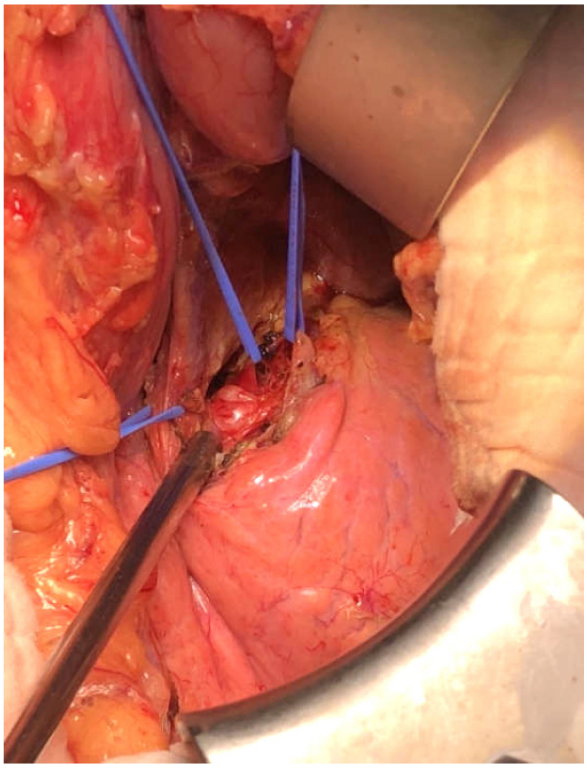

Figure 4 Demonstration of the median arcuate ligament at the level of superior pancreatic border, containing tough fibro-muscular tissues and nerves surrounding the anterior portion of the abdominal aorta and takeoff points of visceral vessels.

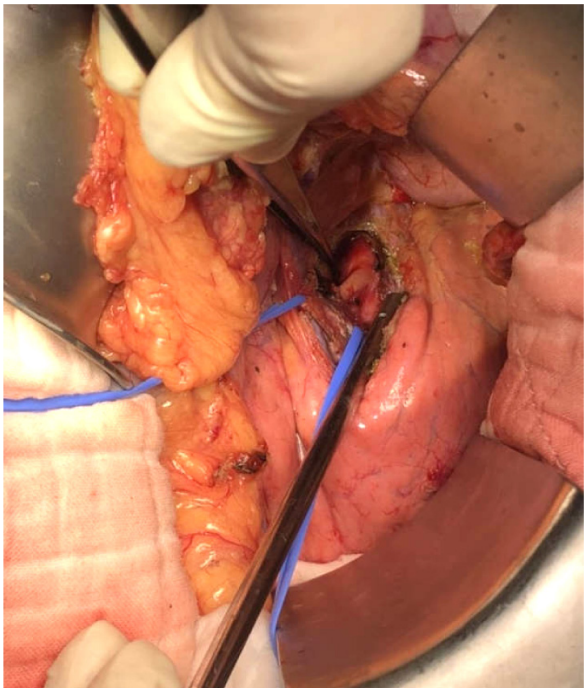

Figure 5 After aggressive resection of the median arcuate ligament and exposing the celiac trunk, the anterior aspect of the abdominal aorta could be seen.

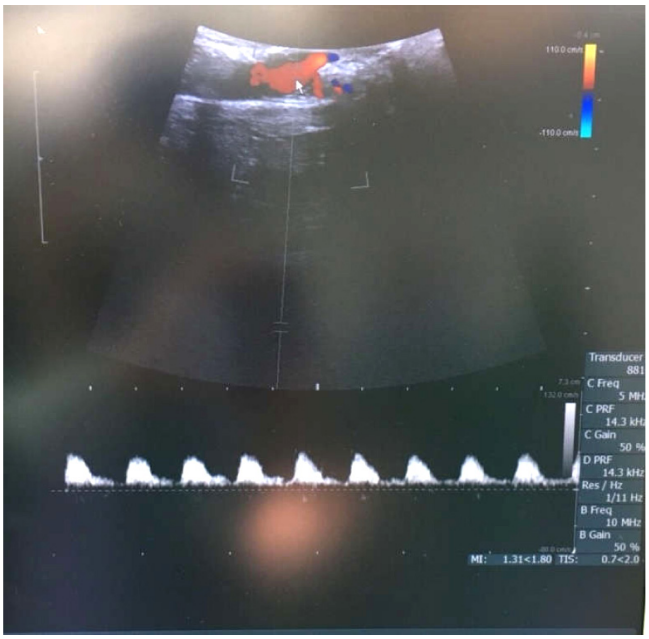

Figure 6 Intra-operative duplex ultrasound showed a celiac artery peak-systolic velocity of 120 cm/sec, verifying adequate decompression.

Discussion

Celiac artery compression syndrome is more prevalent in women compared with men and more common between the ages of 40 to 60 years. The incidence of clinical symptom is much lower than the finding of celiac artery compression from abdominal CT scan[3,4]. The symptoms of CACS most typically occur after meals (94%)[5]. Weight loss is reported in approximately one-half of patients[6,7,8]. In Western countries, the incidence of CACS ranges from 12.5% to 24%, but the incidence is much lower (2.3%) in Japan[9]. Clinical manifestations of CACS included the triad of postprandial pain, nausea and vomiting, and weight loss. It can be easily misdiagnosed as dyspepsia or peptic ulcer disease.

Two main theories are used to explain the symptoms. First is mesenteric ischemia, either foregut ischemia or a postprandial vascular steal from superior mesenteric artery (SMA) collaterals leading to midgut ischemia. Second is neurogenic stimulation, caused by direct compression of the celiac ganglia and plexus leading to splanchnic vasoconstriction or sympathetic pain fiber irritation[10]. Lateral aortography was the gold-standard for the diagnosis of CACS[11], but recently multidetecter CTA has become the most commonly-used modality. Radiologic findings of CACS included thickened MAL, respiratory dependent stenosis of proximal celiac artery with post-stenotic dilatation, and hook-like appearance of celiac artery with indentation of the adjacent aorta[12]. Moreover, duplex ultrasonography can also be used as a screening instrument to diagnose CACS according to Gruber (2012).

Open surgery can be performed via a trans-abdominal or retroperitoneal approach, and the division of the median arcuate ligament and resection of peri-arterial neurofibrotic tissue is usually adequate treatment, although some additional celiac artery reconstruction might improve the outcome. Symptoms were relieved through surgery in 70% to 80% of cases[13]. In 2000, Roayaie *et al*, performed the first laparoscopic treatment for CACS[14] and a case series reported by Buccani *et al*[15] showed equal effectiveness of symptoms relief between open and laparoscopic methods. The advantages of the laparoscopic approach included early oral feeding, shorter hospital stay, faster recovery, decreased post-operative adhesions and the avoidance of morbidity associated with an upper midline laparotomy incision. In 2016, Weber *et al*. reported 39 patients treated with laparoscopic median arcuate ligament resection and found 85% of patients to be symptom-free at follow up, but 10% required conversion to open surgery due to intra-operative hemorrhage[16]. Disadvantages of the laparoscopic approach included the difficulty in dissection and controlling of hemorrhage from the aorta especially if there is fixed stenosis of the celiac artery.

Recently, endovascular therapy of the celiac artery by percutaneous trans-luminal angioplasty (PTA) and stent placement has been reported but the recurrence rate was high and the duration of symptom relief was relatively short. This could be due to continued extrinsic pressure from the surrounding fibromuscular tissues and nerves or median arcuate ligament, causing stent damage. However, in the case of residual stenosis or recurrence of symptoms after surgical celiac decompression, angioplasty with or without stenting can be used if resources are available.

The confirmation of adequate decompression by intra-operative ultrasound demonstrating the return of celiac artery PSV to normal levels does not always predict symptom resolution. This suggests that the pathophysiology of CACS is complex and symptoms might not be related solely to blood flow but perhaps also to the splanchnic nerve plexus. Thus, endovascular treatment of CACS might have limited success[17,18].

Median arcuate ligament and celiac plexus division is adequate treatment for CACS without vascular pathology. Therefore, assessment of flow adequacy must be included in any operation for CACS, as further additional vascular reconstruction might be required. Reilly *et al*. and Takach *et al*.[19] recommended the use of intra-operative duplex ultrasound to confirm normal pressure gradient after decompression. In the present case report, immediate pain relief after operation was seen, and patient could eat normally. At 1 month after discharge, the patient's constipation (a rare presentation) was improved. From a large study, 83% of patients were asymptomatic in the first 6 months after decompression, but this proportion gradually decrease with time. With current limited information, patients with CACS should be followed for long-term re-evaluation after treatment.

Conclusion

CACS is a rare condition, especially in the Orient. Typical clinical characteristics included abdominal pain, epigastric bruit and weight loss, attributed to the compression of the celiac artery and possibly the celiac ganglia by the median arcuate ligament. CTA and conventional angiography are considered to be the gold standard diagnostic modalities. Open surgery is a safe and effective treatment but the choice of treatment also depends on clinical and morphological characteristics of individual patients.

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บทคัดย่อ **รายงานผู้ป่วยที่มาด้วยอาการจากการถูกกดทับหลอดเลือดที่มาเลี้ยงลำไส้ในช่องท้องแขนงที่ 1**

อนุวัชช์ จันทร์ทิพย์

กลุ่มงานศัลยกรรม โรงพยาบาลลำปาง

การกดทับหลอดเลือดที่มาเลี้ยงลำไส้ในช่องท้องแขนงที่ 1 โดยส่วนของกระบังลมและเนื้อเยื่อประสาท ทำให้เกิดอาการปวดท้องหลังรับประทานอาหาร และมีน้ำหนักลดลง เป็นภาวะที่พบได้น้อยและเป็นภาวะที่ต้องทำการตรวจแยกโรคจากภาวะอื่นซึ่งพบได้บ่อยกว่า รายงานผู้ป่วยนี้ได้อธิบายถึงวิธีการตรวจ การผ่าตัดรักษา เพื่อลดอาการของผู้ป่วย